

A Vascular Mimicker: Inferior Vena Cava Leiomyosarcoma Presenting with Pseudo Right Heart Failure and a Massive “Snake Thrombus”

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Abstract

Inferior vena cava (IVC) leiomyosarcomas are exceptionally rare malignancies, accounting for less than 1% of all cancers, and often present as a diagnostic challenge by mimicking right heart failure (RHF). We present a 52-year-old male admitted with progressive dyspnea and generalized edema, where elevated Pro-BNP levels (395 ng/L) initially suggested a primary cardiac etiology. However, the finding of a preserved ejection fraction on echocardiography necessitated further vascular evaluation. Contrast-enhanced CT revealed a 9.0 cm aortocaval mass and a massive Mayo Level IV “snake thrombus” extending into the right atrium. Histopathological examination confirmed IVC leiomyosarcoma. Despite multidisciplinary intervention, the patient’s condition deteriorated rapidly, leading to death within one month of diagnosis. This case highlights the “pseudo-RHF” clinical trap; in patients with volume overload and elevated biomarkers but normal cardiac function, IVC pathologies must be prioritized in the differential diagnosis. Early recognition of such vascular mimickers is vital to prevent life-threatening complications like massive pulmonary embolism and to plan urgent oncological treatment.

Keywords: Inferior vena cava, leiomyosarcoma, right heart failure, tumor thrombus

Introduction

The inferior vena cava (IVC), the largest venous structure in the body, is the main pathway that carries blood from the lower extremities, pelvis, and abdominal organs to the right atrium (1,2). Inferior Vena Cava Syndrome (IVCS), which develops as a result of obstruction or compression of the IVC, is a clinically challenging condition characterized by symptoms such as peripheral edema, ascites, and dyspnea (3,4).

In clinical practice, IVC obstructions create significant difficulties in differential diagnosis because they often mimic the findings of right heart failure (RHF) (1). Signs such as dyspnea, orthopnea, and lower-extremity edema resulting from impaired venous return may be interpreted as favoring a primary cardiac disease (1). Therefore, IVC pathologies should always be excluded in patients who exhibit symptoms of heart failure despite having a normal ejection fraction on echocardiography (1,3,5).

The causes of IVC obstruction span a wide spectrum, ranging from congenital anomalies to acquired pathologies (1). Among the acquired causes, the most common are

malignancy-related tumor thrombi and iatrogenic factors such as filter use (1,4,6). Leiomyosarcomas, which are the primary malignancies of the IVC, are extremely rare tumors accounting for less than 1% of all malignancies (2). These tumors usually progress insidiously and become clinically apparent only when they create a massive intraluminal filling defect or exert compression on surrounding organs (2). Tumor thrombi tend to invade the vessel wall and may sometimes extend into the right atrium, leading to life-threatening mobile masses known as “snake thrombus” (5).

In this case report, we present a 52-year-old man who was admitted with complaints of dyspnea and generalized edema. Because of elevated Pro-BNP level, a cardiac etiology was initially suspected. However, subsequent imaging studies revealed massive thrombosis extending from the right femoral vein to the right atrium, as well as an IVC-derived leiomyosarcoma. This case highlights the critical importance of evaluating the IVC, particularly in patients who present with signs of right-sided heart failure despite having normal cardiac function.

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Case Report

A 52-year-old male patient with no known systemic disease or history of regular medication use presented to our emergency department with complaints of swelling in both lower extremities, pain, nausea, and progressively worsening shortness of breath over the past week. The patient’s history revealed that he had undergone surgery for bilateral inguinal hernia approximately 45 days before presentation. It was reported that the patient was diagnosed with bilateral lower extremity deep vein thrombosis (DVT) 10 days after the operation and was started on low-molecular-weight heparin (LMWH) therapy.

During the initial evaluation in the emergency department, his vital signs were as follows: blood pressure 140/90 mmHg, heart rate 98 beats/minute, body temperature 36.4°C, and oxygen saturation (SpO2) 91% on room air. Physical examination revealed diffuse 3+ pitting pretibial edema in both lower extremities. Marked swelling and edema were observed in the scrotal region. On lung auscultation, minimal rales were heard at the base of the right lung. Abdominal examination revealed tenderness in the epigastric and mid-abdominal regions on deep palpation.

In the laboratory investigations of the patient, whose electrocardiogram (ECG) showed a normal sinus rhythm, elevations were detected in C-reactive protein, transaminases, and Pro-BNP levels. Blood urea nitrogen was found to be mildly elevated, whereas creatinine, creatine kinase (CK), and troponin levels were within the reference ranges (Table 1).

A posteroanterior chest radiograph demonstrated blunting of the right costophrenic angle (consistent with pleural effusion). Scrotal ultrasonography (USG) reported

Table 1: The patient’s laboratory results at the time of admission.

Laboratory tests	Patient result	Reference range
BUN	20.6 mg/dL	7 mg/dL – 20 mg/dL
Creatinine	1.00 mg/dL	0.67 mg/dL – 1.30 mg/dL
CK	81 units/L	50 units/L – 171 units/L
AST	290 units/L	5 units/L – 40 units/L
ALT	153 units/L	5 units/L – 40 units/L
Pro-BNP	395 ng/L	70 ng/L – 133 ng/L
Troponin	<0.010 µg/L	0.01 µg/L – 0.023 µg/L
CRP	9.2 mg/dL	0 mg/dL – 0.8 mg/dL

that “the scrotal skin and subcutaneous tissue appeared markedly edematous and heterogeneous, with increased vascularity observed on color Doppler ultrasonography (CDUS).” Transthoracic echocardiography (TTE) assessed the “left ventricular ejection fraction as normal.”

Contrast-enhanced abdominal computed tomography (CT), performed as part of the etiological investigation, revealed that “a widespread obstructive thrombus is present extending from the right femoral vein through the right external iliac vein, right common iliac vein, and inferior vena cava (IVC) up to the right atrium (Figure 1). At the renal level, there is an aortocaval mass lesion measuring approximately 9.0 cm in diameter (Figure 2), along with diffuse free fluid within the abdomen (Figure 1B).

Following multidisciplinary consultations involving the departments of cardiovascular surgery, general surgery, and oncology, the patient was admitted to the Cardiovascular Surgery Clinic. Histopathological examinations performed after referral to a tertiary healthcare center for further

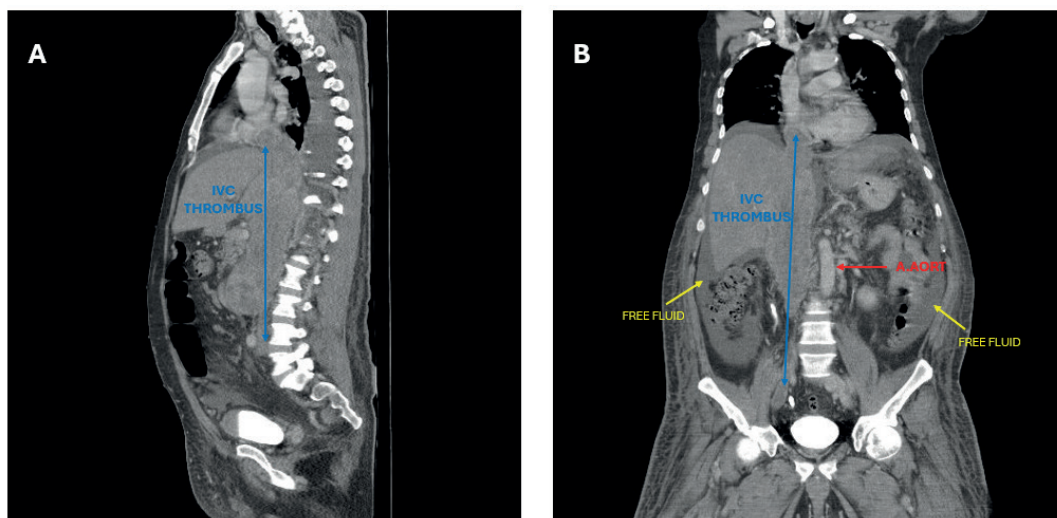


Figure 1. Abdominal Computed Tomography (CT) Findings. (A) On the sagittal section, abnormal wall thickening, mass effect, and thrombus appearance in the inferior vena cava (IVC), showing extensive craniocaudal extension along the posterior abdominal wall. (B) On the coronal section, abnormal wall thickening and mass effect in the inferior vena cava (IVC), the relationship of the thrombus (blue arrow) with the abdominal aorta (red arrow), and the distribution of free intra-abdominal fluid (yellow arrows) are observed.

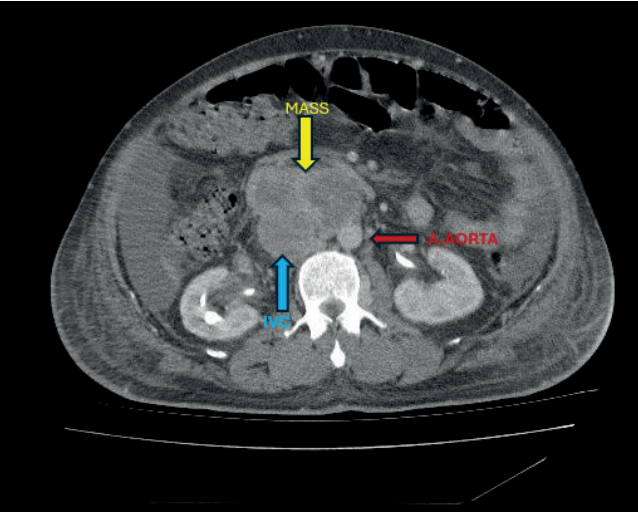


Figure 2. Axial abdominal computed tomography (CT) image of the case. A hypodense/heterogeneous mass (yellow arrow) is seen in the retroperitoneal space, located adjacent to/in front of the abdominal aorta (red arrow) and the inferior vena cava (IVC, blue arrow).

evaluation and management resulted in a diagnosis of leiomyosarcoma. Radiotherapy was planned and initiated at the outside center. As the patient’s clinical condition progressed rapidly and general status deteriorated, the patient was transferred to the palliative intensive care unit and was declared deceased within one month of diagnosis.

Discussion

The clinical presentation of inferior vena cava (IVC) obstruction often poses a diagnostic challenge because it can mimic congestive heart failure (CHF) (1,3). In this case, the patient’s complaints of progressive dyspnea, orthopnea, and bilateral lower extremity edema are classic signs of right-sided heart failure (1). The detection of an elevated Pro-BNP level of 395 ng/L during the initial evaluation complicated the diagnostic process, as this biomarker is typically associated with primary cardiac dysfunction. The finding of a normal ejection fraction on echocardiography highlighted a critical clinical point regarding the need to

exclude Inferior Vena Cava Syndrome (IVCS) in patients who present with symptoms of volume overload but have normal cardiac function.

The pathophysiology of IVCS in this patient is based on reduced venous return (preload) to the heart (1,3,4). A massive thrombus extending to the right atrium and a 9.0 cm aortocaval mass were found to cause near-complete obstruction of the IVC. This obstruction increased systemic venous pressure, leading to the leakage of fluid into the interstitial spaces; clinically, this manifested as +3 pitting edema, scrotal edema, and ascites (diffuse free fluid). Over time, the inability of the right heart to maintain adequate preload results in compensatory responses such as tachycardia and systemic congestion that is indistinguishable from advanced-stage CHF (1). The marked elevation in liver enzymes and the mild increase in BUN reflect secondary organ congestion and dysfunction resulting from impaired venous outflow (4).

The underlying etiology, primary IVC leiomyosarcoma, is an extremely rare smooth muscle tumor accounting for less than 1% of all malignancies (2). These tumors are known for their insidious progression, often remaining asymptomatic until they cause significant obstruction or external compression (2). Extension of the thrombus into the right atrium classifies the condition as a Level IV thrombus according to the “Mayo Clinic Tumor Thrombus Classification” (1,2) (Table 2). Such structures with atrial extension are often referred to as “snake thrombus” because of their high mobility and echocardiographic appearance, and they carry a life-threatening risk of massive pulmonary embolism (5).

During the diagnostic process, distinguishing a tumor thrombus from a bland thrombus is critically important, especially considering the patient’s history of postoperative DVT (1,2). Unlike bland thrombi, tumor thrombi demonstrate contrast enhancement on CT, expansion of the vascular lumen, and direct continuity with the primary mass (1,2,7). In this case, the 9.0 cm mass and the filling defect, which exhibited vessel-expanding and contrast-enhancing characteristics, confirmed the malignant process.

Table 2: Mayo Clinic Tumor Thrombus Classification and TNM Equivalent Correspondence

Level	Classification	Anatomical Definition	TNM Equivalent
Level 0	Renal Vein Only	The tumor thrombus is confined to the main renal vein and has not reached the inferior vena cava (IVC).	T3a
Level I	Lower Inferior Vena Cava	The thrombus extends into the IVC, but its apex is within 2 cm above the renal vein ostium.	T3a
Level II	Mid Inferior Vena Cava	The thrombus extends > 2 cm above the renal vein junction, but its apex remains below the hepatic veins.	T3b
Level III	Upper/Intrahepatic IVC	The thrombus extends into the IVC behind the liver; it is above the hepatic veins but below the diaphragm.	T3b
Level IV	Supradiaphragmatic	The thrombus extends beyond the diaphragm up to the right atrium of the heart.	T3c

This case demonstrates that IVC pathologies should be considered a primary differential diagnosis in patients presenting with prominent signs of right heart failure and elevated Pro-BNP levels but preserved ventricular function on echocardiography. Early diagnosis of a malignant IVC thrombus is of vital importance because of the complexity of its surgical management and its prognosis, which differs from that of primary cardiac diseases.

Conclusion

This case, characterized particularly by widespread edema and dyspnea, demonstrates that IVC obstructions can create a clinical picture similar enough to be mistaken for right heart failure. Despite the elevated Pro-BNP level detected in the patient, the preservation of ejection fraction on echocardiography indicates that the clinical focus should be shifted from primary cardiac dysfunction to a vascular obstruction (Inferior Vena Cava Syndrome). Early diagnosis of rare and invasive pathologies such as a Mayo Level IV thrombus and leiomyosarcoma is a vital necessity for preventing life-threatening pulmonary embolism complications and for planning complex surgical treatment modalities.

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